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Images in Congenital Heart Disease

Late diagnosis of common arterial trunk

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A 61 YEAR OLD FEMALE WAS REFERRED FOR increasing cyanosis, syncope and exercise intolerance of nine months duration. In early childhood, she had been diagnosed with tetralogy of Fallot, and told that her cardiac condition was inoperable. Physical examination revealed a thin, tachypnoeic female with severe cyanosis and clubbing. She had a loud systolic ejection murmur in the right second intercostal space, a loud, single second heart sound, and a parasternal heave. Saturations of oxygen, measured transcutaneously, were 62%. The chest radiograph showed cardiomegaly, and a right aortic arch. On echocardiography, there was large outlet ventricular septal defect with a single arterial, but technically difficult windows obscured morphology of the central pulmonary arteries. Accordingly, cardiac magnetic resonance imaging and post-contrast imaging were performed to elucidate the origin of the flow of blood to the lungs. The steady-state free precession shown in Figures 1 and 2 confirmed a large subarterial ventricular septal defect (VSD), the crest of the ventricular septum being overridden by a common arterial trunk guarded by a common arterial valve, which was regurgitant, as well as visualizing the pulmonary arteries. Three dimensional reconstruction (Figs. 3 and 4) further clarified separate, adjacent origins of the pulmonary arteries from either side of the ascending common trunk. There was a single coronary artery arising from the posterior truncal arterial sinus (Fig. 5). Based on these findings, the final diagnosis was common arterial trunk, with separate origin of the pulmonary arteries from the side of the trunk, right aortic arch with mirror image brachiocephalic branching, trileaflet truncal valve overriding a large interventricular communication, mild truncal valvar regurgitation, and severe pulmonary hypertension.

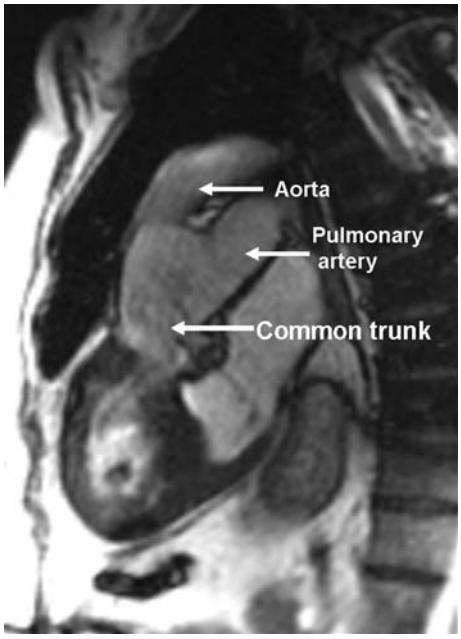

Figure 1.
Fast-gradient-echo steady-state free-precession (SSFP) cine cardiovascular magnetic resonance (CMR) image visualizing the common arterial trunk and large subarterial VSD.

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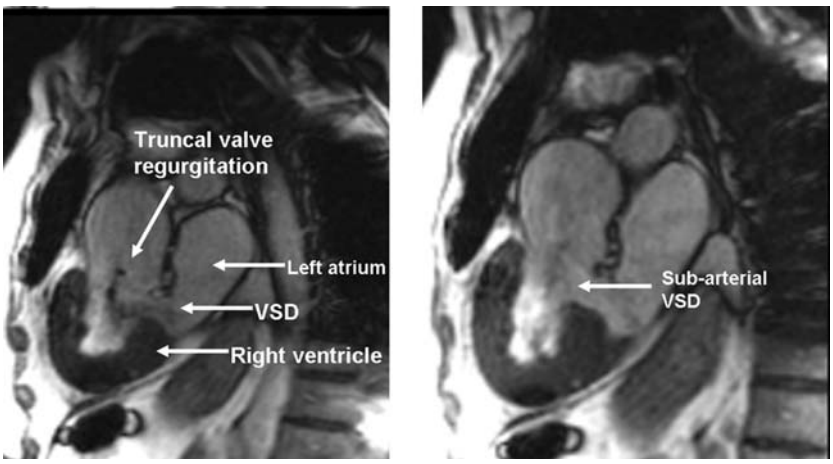

Figure 2.
Oblique SSFP cine CMR images show the severely hypertrophied right ventricle and truncal valve regurgitation.

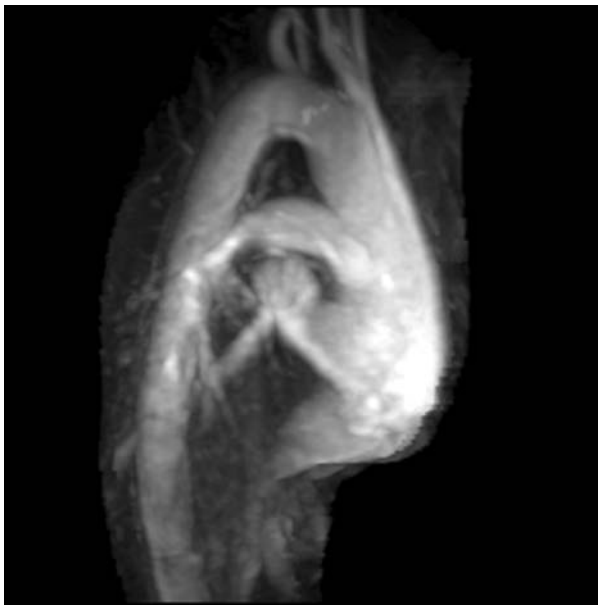

Figure 3.
Three dimensional (3D) reconstruction of gadolinium enhanced magnetic resonance angiography (MRA) visualizing the separate, adjacent origins of the pulmonary arteries from either side of ascending common trunk.

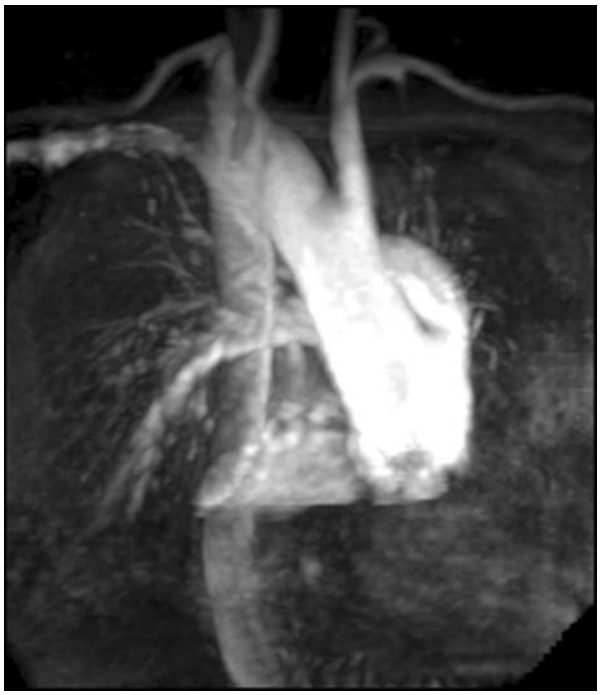

Figure 4.
3D MRA demonstrating the right aortic arch and mirror image branching of brachiocephalic arteries.

Although inoperable, the patient was treated with sildenafil citrate for her pulmonary hypertension, and six months later reported considerable subjective improvement in her symptoms.

The clinical course of patients with common arterial trunk is immensely variable, but most untreated patients die in early infancy or childhood as the result of congestive heart failure and coronary arterial insufficiency from massive flow of blood to the lungs.[1] To our knowledge, our patient is the oldest surviving with untreated common arterial trunk and confluent pulmonary arteries. Cardiac magnetic resonance imaging gave precise definition of the origins, size, and

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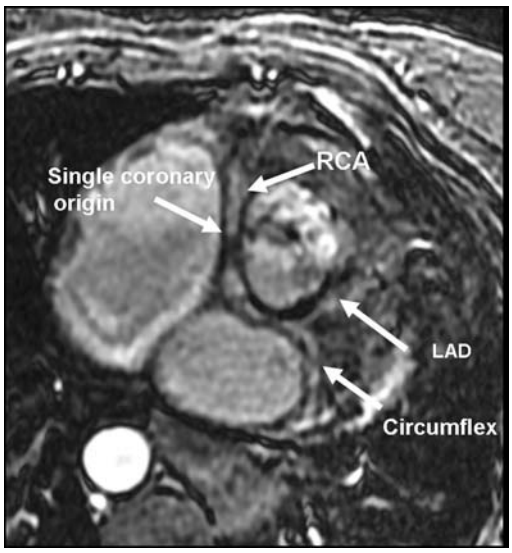

Figure 5.
3D SSFP coronary artery imaging demonstrates a single coronary artery arising from posterior truncal arterial sinus and bifurcating in to right and left coronary arteries.

distribution of the pulmonary arteries and offers an excellent, safe, noninvasive imaging modality in this clinical situation.

Reference

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